Tumors of the vertebral column

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Tumors of the vertebral column, with the exception of bone metastases are uncommon. They represent however an important cause of morbidity and mortality and require clear understanding of their biology and thorough knowledge of the various management alternatives.

Bone tumors represent less than 1% of all cancers; of those, less than 10% are localized in the axial skeleton. Metastases are the most common by far, followed by myeloma, primary bone neoplasms being much rarer.

The vertebral column contains a variety of histological components - osteoid, cartilage, fibrous tissue, vascular elements and bone marrow. All these can give origin to neoplasms. Thus the histological variety sarcomas is remarkable. In addition, paraspinal tumors, particularly sarcomas may secondarily involve the vertebra and it is sometimes not possible to determine their exact site of origin.

In the following table from Dahlin (1978) are indicated the most common malignant tumors of the spine excluding metastases.

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myeloma</td>
<td>34.6%</td>
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<tr>
<td>Chordoma</td>
<td>28.4%</td>
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<tr>
<td>Lymphoma</td>
<td>10.5%</td>
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<tr>
<td>Chondrosarcoma</td>
<td>9.8%</td>
</tr>
<tr>
<td>Osteosarcoma</td>
<td>6.4%</td>
</tr>
<tr>
<td>Ewing's sarcoma</td>
<td>5.0%</td>
</tr>
<tr>
<td>Fibrosarcoma</td>
<td>3.4%</td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>1.4%</td>
</tr>
<tr>
<td>Malignant giant cell tumor</td>
<td>0.5%</td>
</tr>
</tbody>
</table>

The clinical is, in most instances, non-specific. Pain is a common symptom, and may be spontaneous or be of a mechanical nature. It may simulate all the clinical syndromes of the degenerative spinal disorders, and thus it is not surprising that the correct diagnosis is often quite delayed. An expanding mass, which may reach considerable size is another manifestation. Finally as the tumor encroaches upon the spinal canal, it may cause various neurological syndromes due to involvement of the spinal cord or nerve roots.

An attempt to establish a precise pre-operative diagnosis should always be carried out. The age of the patient should always be taken into consideration, since some tumor types such as osteoid osteomas, Ewing's sarcoma or aneurysmal bone cysts are more common in the younger age groups, whereas metastatic bone disease or multiple myeloma are seen usually in more advanced ages.

Radiographic studies are most helpful and should always be carefully analyzed. In most cases, a variable degree of bone destruction is present, and the more malignant the tumor, the larger the erosion. Less aggressive neoplasms will have a geographical pattern, with a well defined outer margin, whereas malignant tumors will have a rather poor demarcation. Calcification is present in many instances. In tumors of cartilaginous origin the calcifications are frequently central and are flocculent or flecklike. It is also observed in the tumor matrix of osteomas or osteosarcomas. Vertical striation is typical of vertebral hemangiomas. Periosteal ossification in concentric layers with an onion peel pattern is characteristic of Ewing's sarcoma and osteogenic sarcomas.

CT scan of the spine and Magnetic Resonance are indispensable to evaluate the degree of bone destruction, to outline the compromise of the various vertebral components, to appraise the degree of compression of the neural structures and the extension of the soft component.

It is sometimes difficult to decide whether spine angiography should also be carried out. We feel that if one suspects the presence of a vascular lesion - aneurysmal bone cyst, hemangiopericytoma, hemangioma - or if one wants to define vascular anatomy of the cord, a detailed vascular study is strongly advisable. In addition, pre-operative embolization is extremely helpful in dealing with vascular tumors.

Apropriate laboratory studies should also be
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Isotope scans with Technetium may be quite helpful to map the extent of bony involvement when multiple lesions are present or when there is trouble to localize a suspected lesion, such as an osteoid osteoma.

As far as the treatment is concerned a few points should be kept in mind. The primary goals may vary from case to case, but in the presence of a neurological deficit decompensation of the neural structures is the main purpose. The appropriate surgical route (anterior, posterior, posterolateral) depends on the anatomical features of each case, and the segment of the spine involved.

In each case, a specimen for histological diagnosis has to be procured. The method chosen should not jeopardize a more radical or definitive treatment. In cases such as lymphomas or eosinophilic granulomas obtaining a small piece of tissue will be enough. In contrast, one should attempt to remove radically an osteoid osteoma, or to «clean» as much as possible an aneurysmal bone cyst.

It is also important to emphasize that the bone lesion itself may be cause of spinal instability, and this can be aggravated by the surgical procedure. It is therefore quite important in these circumstances to stabilize the spine, either with the help of metallic prosthesis, bone grafting or bone cement. Injection of the vertebral body with acrylic has been proposed in some vascular tumors such as hemangiomas. Once again it should be emphasized the adjuvant role of embolization in some vascular lesions.

Surgery alone is certainly not the answer for some of these neoplasms. Therefore a multidisciplinary approach including orthopedic surgeons, radiotherapists and oncologists is mandatory. Ewing's sarcoma, osteosarcomas, multiple myeloma are paradigms of lesions which can be dealt only by the cooperative effort of experts in these various fields.

Osteoid osteoma

Represents 3.17% of primary bone tumors and 1.37% of the tumors of the vertebral column. These are benign osteoblastic lesions with less than 1cm in diameter. They affect more often young males, and 50% are localized to the pedicle or lamina. Pain is a common complaint, and is worse at night or by motion, and in 25-40% of the cases is relieved by aspirin. Local tenderness, scoliosis and radicular symptoms are the other manifestation. Radiographically they show a central radiolucent nidus, with a surrounding density, but they may be quite difficult to detect. Isotope bone scan is then extremely helpful. Complete excision of the lesion is usually curative.

Osteoblastoma

This is an osteoblastic tumor with more than 1 cm of diameter, often with expansion into the spinal canal. Radiographically they present as lytic lesions with variable degrees of central ossification and periosteal bone formation. A complete surgical removal should be attempted.

The role of the radiotherapy is controversial.

Hemangioma

This is a benign lesion present at autopsy in about 8.9-12.5% of the cases. They are single or multiple and usually asymptomatic. They may be associated with the metameric cutaneous lesions -Klippel-Trenaunay-Webber syndrome. They may cause neurological manifestations by subperiosteal growth, by the development of an epidural mass, by collapse (pathological fracture), by expansion of the posterior rim, widening of the pedicles and laminae, or by hemorrhage. Some of these lesions may become symptomatic during pregnancy. Despite their usually characteristic radiographic appearance, with the typical vertical striations the differential diagnosis is to be made with metastatic disease. They are extremely vascular and if a surgical decompression is to be carried out pre-operative embolization is quite helpful.

Giant cell tumor

Also called osteoclastomas are rare lesions in the spine. They cause an area of rarefaction, expanding the body or the arch, sometimes with a multiloculated appearance giving it the aspect of «soap-bubble». They are more common in the second or third decade and have a predilection for females. Surgical excision is recommended, but there is a high risk of recurrence (about 50%) and a 10% malignant evolution.

Aneurysmal bone cysts

These are benign lesions, appearing in children and young adults characterized by an expansile erosion with
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a thin of bone, involving sometimes multiple segments. Some are quite vascular and pre-operative embolization may be helpful. Curettage or excision are advisable.

Benign chondrogenic tumors

These comprise a number of histological types. Chondromas are made of mature hyaline cartilage, and are located in the center of the bone tissue - «enchondromas» or subperiosteally - «periosteal chondromas». They are quite rare in the spinal column.

Osteochondromas are more common, and constitute 46.2% of all benign bone tumors. When multiple, they are part of an hereditary disorder called «multiple osteochondromatosis» or «multiple hereditary exostosis». They begin in early childhood, from within the periosteum and grow by endochondral calcification of the cartilage cap. They may impinge upon the canal and require surgical excision.

Benign chondroblastoma and chondromyxoid fibromas are quite rare, the latter developing from connective tissue forming cartilage.

Eosinophilic granuloma

These may be solitary or polyostotic and affect usually children and young adults. They cause circumscribed areas of osteolysis, and are a common cause of vertebral plana in immature spines. They may be considerable vertebral collapse without neurological deficit. Low dose radiotherapy may be indicated and frequently there is reconstitution of the vertebral body.

Chordoma

Represents 1-4% of all malignant bone tumors. These tumors originate from the embryonic notochord. 55% are located in the sacrococcygeal region, 35% in the clivus and 15% are vertebral. They are most frequent in males and the peak of incidence is around 50-70 years. In the sacral region they often present with pain and alteration of the bowel habits and may be detected by a simple rectal examination. In the cervical area they may grow into the retropharyngeal space and cause dysphagia. They can reach considerable size in the pelvic area. They are lobulated, often with a pseudocapsule, greyish, solid, with areas of cystic degeneration, calcification or mucoid appearance. The characteristic cell types are the «physaliphorous» cells with ample, vacuolated cytoplasm, and the «signet-ring» cells. They may present a chondroid transformation and this seems to be associated with a more benign clinical course. Metastases to the lung, bone, soft tissues or liver, have been described in 28-40% of the cases.

Surgery is the treatment of choice, and may be curative in a small number of cases, particularly when the tumor is located in the lower sacral segments. Radical excision in lesions affecting the S1 and S2 segments is associated with major morbidity. Multiple palliative procedures are sometimes worthwhile. Palliative radiation is also advisable. Recently the use of «Bragg peak» modalities has been suggested, as this allows the deliver of higher doses of radiation, with rather encouraging results.

Osteogenic sarcoma

Primary osteogenic sarcoma of the spine is quite rare - 8.5 to 2% of all osteogenic sarcomas. The mean age of incidence is 35, and there is a higher incidence in males. Paget's disease and previous irradiation are predisposing conditions.

These tumors are more common in the lower segments. They are characterized by the production of osteoid by the stroma. Fibroblastic, chondroblastic, osteoblastic and mixed varieties have been described. Radiographically they are dense lesions, often with invasion of the soft tissues. Radical surgical removals associates with multimodality chemotherapy have improved the outlook of these patients.

Chondrosarcoma

Represents 10-20% of all malignant bone tumors, second only to osteosarcomas. They may arise «de novo» or be secondary to other diseases such as multiple osteochondromatosis, in which malignant degeneration is seen in 10% of the cases. They are most common in the pelvis extending subsequently to the sacrum and in the head of the ribs growing secondarily to the vertebral canal. Radiographically they present as radiolucent destructive lesions often containing areas of speckled calcifications. The mesenchymal chondrosarcoma is a most malignant variety. Surgical excision and chemotherapy are indicated.
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Myeloma

These are the most common primary bone tumors, and they originate from the proliferation of B lymphocytes. The disease may affect multiple bones or, in about 3% of the cases, be localized - solitary plasmacytoma. In 40% of these, the process is localized to the spine. The vertebral body is involved in most instances but the lesion may extend to the pedicles and laminae and have a large soft tissue component. Neurological syndromes may result from diffuse osteoporosis, compression fractures, or involvement of the spine cord and roots. The solitary form is more common in males, occurs in the fifth decade and in only 30% of the cases there is the characteristic M-protein (whereas this is present in about 90% of the multiple myeloma). The majority however will become generalized. The prognosis is better for the solitary tumor (median survival 10 years) than for the multiple myeloma (median survival 2 years).

One should suspect of this lesion in the presence of severe, protracted pain, weight loss, malaise, anemia and other systemic symptoms. Appropriate laboratory work up is mandatory trying to characterize the paraproteins secreted by the tumor cells.

Once the diagnosis is made careful planning of the therapeutic strategies is indispensable. Decompression and stabilization of the bone spinal framework should be performed due to the risk of collapse during subsequent chemotherapy and radiotherapy. Appropriate decompression is also necessary in patients with disabling radicular pain syndromes.

Ewing’s sarcoma

This neoplasm of probable mesenchymal origin, represents about 5-10% of the primary bone lesions. In 5% of the cases is located in the spine and in 50% in the pelvis or sacrum. It affects people in the 2nd and 3rd decade. It is now considered a systemic neoplasm and at the time of diagnosis is already widespread in 25 to 50% of the cases. It causes lysis of the bone and may reach a large volume. Histologically it is made of small cells, is quite vascular and contains areas of necrosis and no osteoid tissue. Biopsy is mandatory for the correct diagnosis, and this should be followed by chemotherapy before a major surgical resection is undertaken.

Lymphoma

These are usually of the non-Hodgkin type and may be primary - the old «reticulum cell sarcoma» - or secondary. They have a peak of incidence in the second decade and cause destruction of the bone, sometimes a large multifocal lytic lesions, associated with a soft tissue component. Excellent results may sometimes be expected with local radiotherapy associates or not with chemotherapy.

Hemangiopericytoma

Involvement of the spine by these vascular tumors is uncommon. They originate from the pericyte a cell localized around the reticular sheath of capillaries and postcapillary venules. They are quite vascular, and metastases are seen in 15% of the cases. Surgical excision is the treatment of choice, and should be preceded by embolization of the lesion.

Malignant fibrous histiocytoma and fibrosarcoma

These are very rare lesions originated from the histiocytros, and causing destruction of the bone tissue. Chemotherapy is indicated.